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A Case of Bilateral Lambdoid and Sagittal Synostosis Diagnosed with Skull Fracture after Vacuum-assisted Delivery: A Case Report and Literature Review

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Abstract

Craniosynostosis (CS) can develop in the fetal period, but it is difficult to diagnose prenatally. In this case, a 3-month-old female baby developed extensive subgaleal hematoma and severe anemia after vacuum-assisted delivery. Her computed tomography showed bilateral lambdoid and sagittal synostosis (BLSS) with a depressed fracture of the right parietal bone. She was referred to our hospital for treatment of the CS. At 4 months of age, she underwent bilateral lambda and sagittal suturectomy and foramen magnum decompression.

CS may result in trauma at delivery, because CS disturbs fetal head molding during delivery and disrupts passage through the birth canal. In particular, the risk of severe peripartum trauma is thought to increase in cases of CS with multiple suture fusions, such as those observed in BLSS, due to the strong inhibition of this process of passage through the birth canal. Therefore, if the delivery is abnormally prolonged or if the infant has a massive subgaleal hematoma, it is important to perform evaluations for CS after birth.

Keywords: craniosynostosis, bilateral lambdoid and sagittal synostosis, fetal head molding, skull fracture, maternofetal trauma

Introduction

Craniosynostosis (CS) can develop in the fetal period. It has been reported that undiagnosed prenatally developed CS may be a risk factor for delivery trauma and may be associated with cephalopelvic disproportion and abnormal delivery outcomes.[1,2] It is desirable to have CS diagnosed prenatally for safe delivery, but this may be difficult to achieve during the fetal period. In this article, we report on and review literature related to a case of CS diagnosed following a subgaleal hematoma and skull fracture after aspiration delivery.

Case Report

The patient was a female baby with no noted fetal abnormalities. She was delivered by vacuum extraction at 38 weeks and 6 days at another hospital. She was born with an Apgar score of 7/8, weight of 3308 g, height of 52.5 cm, head circumference of 34.8 cm, and delivery time of 9 h 15 min. Blood tests revealed no abnormalities in the coagulation system, such as hemophilia. A computed tomography (CT) scan of the head was performed on day 1 after birth due to extensive subgaleal hematoma and severe anemia. The CT image showed extensive subgaleal hematoma throughout the head without intracranial hemorrhage (Fig. 1a, b). A three-dimensional (3D) CT image revealed bilateral lambdoid and sagittal synostosis (BLSS) and a de-

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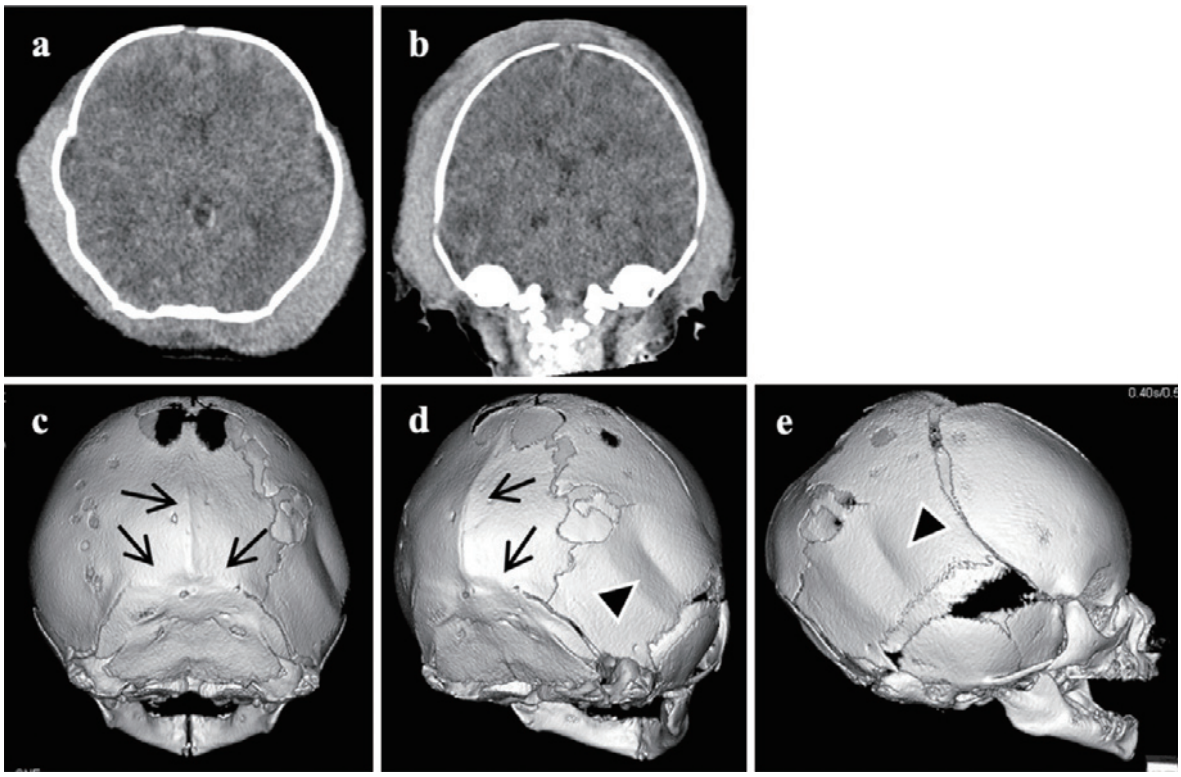

Fig. 1 Computed tomography images of the head at day 1 after birth.
(a: axial, b: coronal) Extensive subgaleal hematoma is seen throughout the head computed tomography scan, but there is no intracranial lesion.
(c, d, e: 3D) A depressed fracture of the right parietal bone from the temporal to parietal direction (arrow head) and bilateral lambdoid sagittal synostosis (black arrow) was noted.

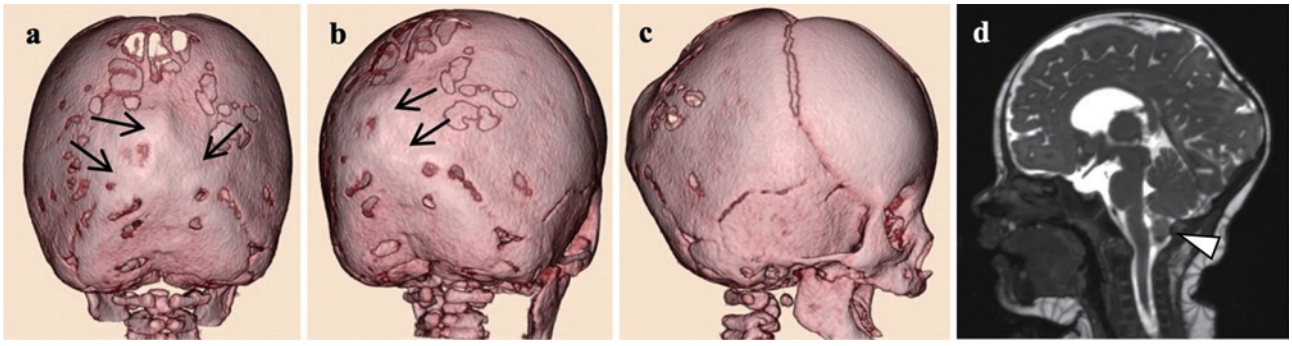

Fig. 2 Computed tomography and magnetic resonance images of the head at 3 months of age.
(a, b, c) Bilateral lambdoid sagittal synostosis (black arrow) had progressed, but the depressed fracture had healed.
(d) Mild cerebellar tonsillar herniation (white arrow head) was observed.

pressed fracture of the right parietal bone (Fig. 1c, d, e). She was discharged without complications at 27 days of age after partial exchange transfusion for severe anemia and phototherapy for jaundice and was later referred to our hospital at 3 months for treatment of the CS. A follow-up CT scan at our hospital showed progressive fusion of the lambda-sagittal suture (Fig. 2a, b), but the depressed fracture had healed spontaneously (Fig. 2c). An MRI at 3 months also showed mild cerebellar tonsillar herniation (Fig. 2d).

At 4 months of age and a weight of 6.4 kg, the patient underwent bilateral lambda and sagittal suturectomy and foramen magnum decompression with a coronal skin incision (Fig. 3a, b, c). The operative time was 3 h 41 min, blood loss was 65 mL, and 86 mL of red cell concentrate was transfused. Due to heavy bleeding from the dura

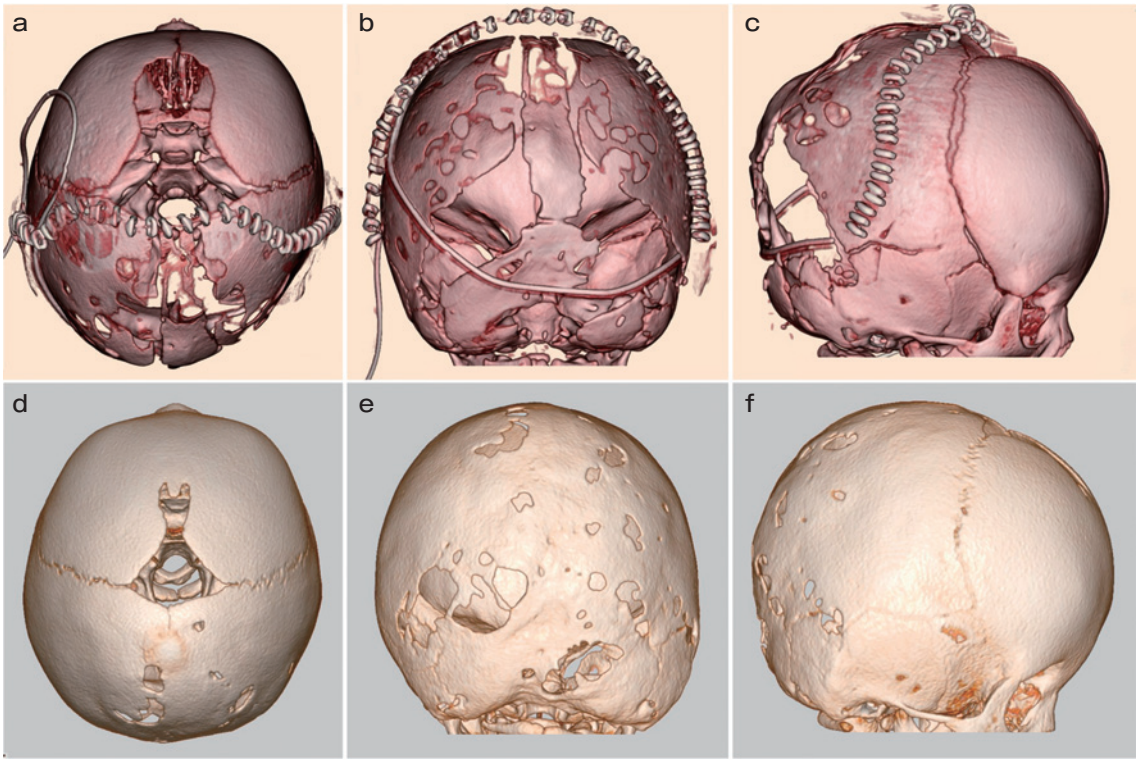

Fig. 3 Postoperative three-dimensional computed tomography.
(a, b, c) The day after surgery.
In addition to foramen magnum decompression from the right side, due to heavy bleeding from the dura around the superior sagittal sinus, we performed a suturectomy excluding the posterior portion of the sagittal suture (d, e, f). Approximately 2 years and 1 month after surgery, cranial morphology has improved, but bilateral lambda and sagittal sutures tend to be fused.

around the superior sagittal sinus, we performed the suturectomy excluding the posterior portion of the sagittal suture. In addition, a foramen magnum decompression from the right side was also undertaken. The patient had a good postoperative course and was discharged without complications 9 days after surgery. Helmet therapy was initiated on the 17th postoperative day, and she has a good developmental quotient index with 99 on the Kyoto Scale of Psychological Development at 1 year and 7 months, which is within normal limits. She is now 2 years and 5 months old and presents with good cranial morphology, but the sagittal and lambda sutures are tending to fuse (Fig. 3d, e, f).

Discussion

BLSS was first described by Neuhauser et al. in 1976 and is also known Mercedes-Benz syndrome due to the posterior view of the fusion morphology.[3] It includes brachycephalic and dolichocephalic morphologies, and the head shape observed in BLSS depends on the prevalence of prematurely fused sagittal and/or lambdoid sutures. Intracranial lesions include cerebellar tonsillar herniation with narrowing of the posterior cranial fossa, venous malformations, and ventricular enlargement. Treatment strategies differ according to the phenotype, with dolichocephaly being mainly treated by sagittal suture resection to reduce the anterior-posterior diameter and brachycephaly being treated by decompression of the narrowed posterior cranium.[4-6] Because the patient in our case was classified as brachycephalic and has a narrowed posterior cranial fossa and increased intracranial pressure, suture resection and foramen magnum decompression were chosen for treatment.

Although CS is typically diagnosed through the observation of cranial deformities in infancy, in some syndromic CS cases, it can be diagnosed prenatally as early as 19 weeks, using ultrasonography.[7] Syndromic CS can be diagnosed by visualizing abnormalities in other parts of the body such as morphological abnormalities of the face, trunk, and fingers. In contrast, the prenatal diagnosis of non-syndromic CS is difficult because the timing and extent of fusion vary from case to case, and non-syndromic CS has no other systemic malformations. Nevertheless, there has been active research on the prenatal diagnosis of non-syndromic CS using ultrasonography, and diagnostic accuracy is improving. Prenatally diagnosed cases of non-syndromic CS have been reported in the literature.[8,9] A

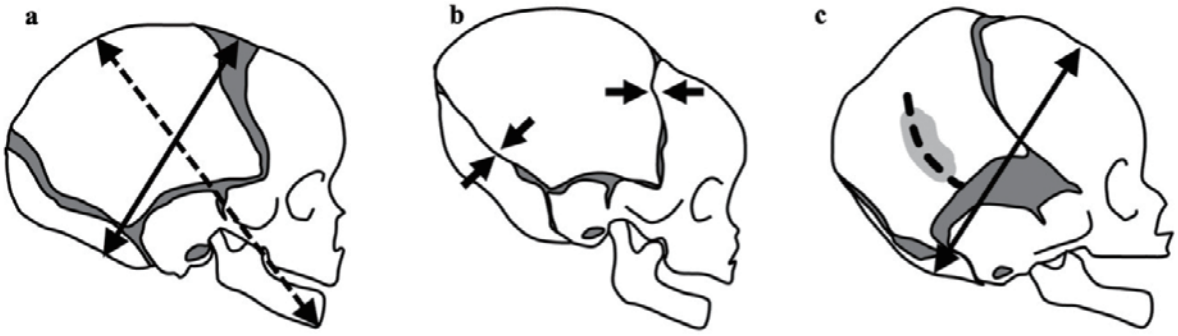

Fig. 4 Fetal head molding.
(a) Normal fetal craniofacial formation.
Solid line: suboccipito-bregmatic diameter, dotted line: mento-vertical diameter.
(b) Molding makes the frontal and occipital bones move inward and the sutures close. The suboccipito-bregmatic diameter decreases, and mento-vertical diameter increases.
(c) Fracture site.
The fracture line (dotted line) extended in the direction causing a shortening of the suboccipito-bregmatic diameter resulting in depression of the parietal bone.

prenatal genetic diagnosis of syndromic CS is possible by amniocentesis or chorionic villus sampling if the causative gene has been identified. However, this is impossible for non-syndromic CS. MRI cannot visualize cranial sutures and is impractical to perform as a screening test in all cases. Therefore, there is currently no established, valid alternative test.[10] Several studies have reported that cranial suture ultrasonography is useful in postnatal diagnosis[11] and fetal ultrasonography is expected to be used in the future. It remains unknown whether there are differences in the clinical course of cranial deformation based on the timing of onset of CS; however, cranial deformities tend to be severe in cases detected early. Regarding the timing of treatment, less-invasive endoscopic suturectomy following cranial remodeling orthoses can be performed if the diagnosis is made during the fetal period.[12]

Head injuries during delivery include extracranial injuries such as cephalohematomas, subgaleal hematomas, skull fractures, and intracranial hemorrhages such as subdural hematomas, superficial parenchymal, and leptomeningeal hemorrhages.[13,14] Risk factors for cranial trauma during delivery include instrumental delivery, such as vacuum and forceps deliveries; abnormalities related to the delivery process, such as abnormal fetal position and rotation; and maternal-fetal factors such as cephalopelvic imbalance and CS.[15-17] CS is thought to inhibit bone overlap, which delays delivery and results in such peripartum trauma such as cephalohematoma or subgaleal hematoma (Supplementary Table 1).[1,2,17] However, to the best of our knowledge, there have been no reports of skull fractures or intracranial hemorrhages in patients with CS. In the present case, there was no intracranial hemorrhage or parenchymal brain injury, but there was extracranial soft tissue injury and a depressed skull fracture that differed in morphology from a ping-pong fracture.

Fetal cranial morphological changes during delivery have been described to occur through a mechanism known as fetal head molding (FHM).[18] The fetal skull bones are superimposed at the anterior and posterior fontanelles owing to external pressure during delivery. In addition, the parietal bones are deformed by stretching and straightening, resulting in shorter suboccipito-bregmatic diameters and longer mento-vertical diameters, allowing the cranial morphology to adapt to passage through the birth canal (Fig. 4 a, b). The problem with CS during this FHM is that suture fusion prevents bone overlap and changes in morphology. We hypothesize that the obstructive passage through the birth canal due to disruption of FHM resulted in the need for excessive aspiration during delivery, leading to an extensive subgaleal hematoma that required blood transfusion and skull fracture. It is thought that the multiple suture fusions and the closure of the posterior fontanel seen in BLSS strongly inhibited FHM, resulting in the observed fracture. This observation is also supported by the fact that the depressed fracture occurred not as a typical linear or circular ping-pong fracture in this case but in a direction that caused the suboccipito-bregmatic diameter to shrink (Fig. 4c). However, no similar fractures have been reported to date, and this is merely a hypothesis. In this case, the fetus was slightly larger with a body weight of 3308 g and a head circumference of 34.8 cm, suggesting the possibility of cephalopelvic disproportion. Furthermore, the CT scan at birth already showed strong fusion in the lambda sutures and the posterior parts of the sagittal suture were already strongly fused, suggesting that the FHM was extremely difficult to function. We believe that these two factors may be the unique aspects and causes of severe trauma in this case. Although we do not believe that a cesarean section should be chosen in all CS cases, in the case of CS that include multiple suture fusions, such as

BLSS or sagittal synostosis, the risk during delivery may be higher. Therefore, if the delivery is abnormally prolonged, or if the infant has a massive subgaleal hematoma, it is important to perform evaluations for CS after birth.

Conclusion

We reported on a case of BLSS discovered as a result of a subgaleal hematoma and skull fracture after vacuum-assisted delivery. FHM did not occur properly due to the BLSS, leading to severe head trauma. Because fetal onset of multiple craniosynostosis can be a risk of head traumas, such as bone fractures during delivery, evaluation for CS is recommended when the delivery abnormality is severe.

Supplementary Material

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Informed Consent

Written informed consent for publication of this case report and accompanying images was obtained from the patient.

Conflicts of Interest Disclosure

All authors declare no conflict of interest. No funding provided.

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